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Case Report

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A rare disease: primary bone lymphoma case report

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ABSTRACT

Primary bone lymphoma (PBL) is a rare subtype of non-Hodgkin lymphoma, comprising a small proportion of primary bone malignancies and extranodal lymphomas. It typically presents with nonspecific symptoms such as localized bone pain, leading to delayed diagnosis. In this case report, we present a 39-year-old male with no prior comorbidities who applied with right hip pain and was ultimately diagnosed with diffuse large B-cell lymphoma of the femur, germinal center phenotype. Radiological findings revealed a large intramedullary lesion with cortical destruction, and diagnosis was confirmed histopathologically. Despite initial treatment with R-CHOP chemotherapy, the disease showed partial resistance, necessitating second-line R-ICE therapy and localized radiotherapy. Post-treatment imaging demonstrated regression of destructive findings, and PET-CT showed minimal residual activity. The patient has remained symptom-free and under observation since 2020. This case emphasizes the importance of a multidisciplinary approach, combining imaging, histopathology, and multimodal therapy in the effective management of PBL, and contributed to the growing body of evidence guiding the prognosis and therapeutic strategy of this rare entity.

Keywords: Primary bone lymphoma, DLBCL, B Cells, germinal center, R-CHOP, FDG-PET

INTRODUCTION

Primary bone lymphoma (PBL) is defined by the World Health Organization (WHO) as one or more bone lesions without distant visceral or lymph node involvement, or as a single bone lesion with or without regional lymph node involvement. PBL is an uncommon subtype of lymphoma. One to three percent of non-Hodgkin lymphomas, five percent of extranodal non-Hodgkin lymphomas, and three percent of primary bone malignancies are PBLs.[1]

While PBL can affect anyone at any age, males are more likely than women to be diagnosed with it between the ages of 45 and 60.[2] Among the most typical clinical signs are palpable mass, soft tissue edema, pathological fracture, and localized bone pain. Nonspecific clinical signs frequently cause a delay in diagnosis; the disease is diagnosed using a mix of imaging investigations and clinical examination, with histological and immunohistochemical examination serving as confirmation. While PBL can arise in any region of the skeleton, the femur, humerus, tibia, spine, and pelvis are the most frequently affected. PBL imaging results are quite erratic and nonspecific.[3] The majority of PBL instances are particularly derived from germinal center centrocytes and fall within the B-cell-like subtype of germinal center cell origin. Based on the existence of miRNA with a particular prognosis, histogenesis, gene expression, and mutation profile, PBL is acknowledged as a unique clinical entity. The prognosis for PBL is good, particularly when combination chemoradiotherapy is used.[4]

In our case, we wanted to examine such cases through a patient diagnosed with PBL, to enable a better understanding of this rare disease and to contribute to the literature.

CASE

A 39-year-old male patient with no known disease applied to the physical therapy and rehabilitation outpatient clinic with a complaint of pain in the right hip. MRI scans were performed. On MRI scans a lesion was present extending along an approximately 20 cm craniocaudal segment, starting from the proximal diaphyseal section of the right femur to the distal diaphyseal section, located intramedullarily. The lesion appeared markedly hypointense on T1-weighted sequences and hyperintense on T2-weighted sequences, demonstrating intense contrast enhancement on post-intravenous contrast sequences. Along the lesion tract, proximally extending approximately 12 cm, there was a solid, well-defined, contrast-enhancing component surrounding the femur, more prominently anteriorly, measuring up to approximately 20 mm in thickness at its thickest point on

contrast-enhanced series. In the proximal diaphyseal section of the femur, limited areas of significant cortical destruction were observed anteriorly. A bone biopsy was taken from the lesion on the MRI. Histopathological examination confirmed the diagnosis of diffuse large B-cell lymphoma with high Ki-67 index, LCA, CD20, BCL-6 and vimentin positive, germinal center phenotype. BCL-2 was negative in bone marrow biopsy, while c-myc and bcl-2 were not tested in the lesion biopsy. No lymph node involvement was detected as a result of PET-CT taken at the time of diagnosis. Bone marrow involvement was not detected in the patient who underwent bone marrow biopsy. In addition, the patient was started on the R-CHOP regimen as chemotherapy. Since the desired treatment response was not obtained in the MRI taken after 6 cycles of R-CHOP chemotherapy regimen, the patient was given 3 cycles of R-ICE chemotherapy regimen as second-line therapy. Curative radiotherapy was applied to the pelvic area for 24 days along with chemotherapy. As a result of the MRI taken for the response to the 2nd line treatment, the destructive findings disappeared and the subsequent PET-CT showed that "an intramedullary lesion showing slightly increased FDG uptake was observed in the proximal right femur (SUVmax: 2.81)", so it was decided to follow the patient without treatment. (**Figure**). The patient continues his regular follow-up without treatment (**Table**). The patient has been symptom-free since 2020, and his findings have been under follow-up (**Table**).

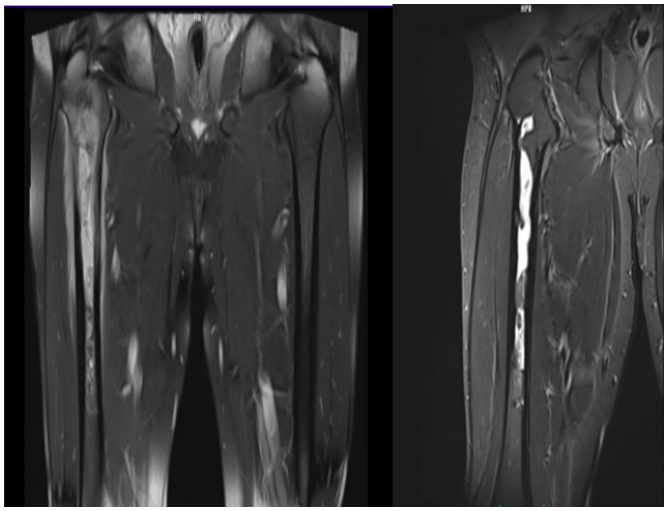

Figure. Femur MRI images of the patient before and after treatment

DISCUSSION

The precise and universally accepted definition of PBL remains a subject of ongoing debate within the medical community. While the disease has been increasingly recognized, there is still no consensus on a standard risk stratification model that accurately predicts the likelihood of its development. Existing scoring systems, although useful in broader oncological contexts, fail to address the specific features and biological behavior of PBL. As a result, clinicians often rely on a combination of clinical judgment and limited prognostic tools when managing these cases.

Among the prognostic models historically employed, the International Prognostic Index (IPI) has been one of the most widely used. However, its utility in the context of PBL has proven to be restricted. This is primarily due to the distinct clinical presentation and progression pattern of PBL compared to other forms of non-Hodgkin lymphomas. In light of this, alternative markers have been explored to better assess disease prognosis and guide therapeutic decisions.

The International Extranodal Lymphoma Study Group (IELSG) 14 study has provided valuable insights in this regard. Notably, the study highlighted the potential prognostic significance of elevated lactate dehydrogenase (LDH) levels in patients with PBL. An increase in LDH may reflect a higher tumor burden or more aggressive disease behavior, thus offering a practical biochemical marker for clinicians in evaluating disease progression. Additionally, several case series in the literature underscore the potential role of radiotherapy as a critical component in the treatment regimen for PBL, often in combination with chemotherapy.[5]

In our own clinical experience, we observed that both LDH levels and radiotherapy played pivotal roles in patient management. Monitoring LDH levels throughout the course of the disease provided important information about the patient's response to treatment and overall prognosis. Furthermore, the inclusion of radiotherapy in the treatment protocol contributed significantly to local disease control, emphasizing its therapeutic value. These observations support the growing body of evidence advocating for a more individualized approach to PBL management, incorporating both biochemical markers and multimodal therapy strategies.

Our findings can be compared with the report by Nishida et al.,[5] who described a case of primary isolated bone marrow diffuse large B-cell lymphoma achieving long-term complete remission with R-CHOP chemotherapy. While our case required second-line therapy and radiotherapy to achieve disease control, both reports highlight the potential for durable remission in PBL with multimodal treatment. This comparison emphasizes the variability in treatment response and the importance of individualized management strategies.

Table. Laboratory values of the patient during treatment and follow-up

	Date	ESR	Hemoglobin (gr/dl)	WBC (10^3/µL)	Platelet (10^3/µL)	LDH (U/l)
At the time of diagnosis	17.04.2019	19	15.3	6.3	260	685
After 1. line of chemotherapy	06.08.2019		12.9	7.1	247	320
After 2. line of chemotherapy	16.12.2019	32	12.6	5.7	45	355
Latest values	21.05.2025	17	15.2	7.07	238	212

ESR: Erythrocyte sedimentation rate, WBC: White blood cell, LDH: Lactate dehydrogenase

CONCLUSION

As a result, PBL represents a rare yet clinically significant subtype of non-Hodgkin lymphoma, characterized by its distinct presentation, biological behavior, and imaging challenges. Despite its low incidence, timely diagnosis and effective management are critical to improving patient outcomes. Our case study reinforces the importance of recognizing key biochemical markers, such as elevated LDH levels, in disease monitoring and highlights the therapeutic value of radiotherapy as part of a multimodal treatment approach. By documenting and analyzing individual cases like ours, we hope to contribute to the growing body of knowledge on PBL and support the development of more refined diagnostic and prognostic strategies for PBL. Further multicentric case series are essential to optimize diagnostic and therapeutic algorithms for this uncommon malignancy.

ETHICAL DECLARATIONS

Informed Consent
The patient signed and free and informed consent form.

Referee Evaluation Process
Externally peer-reviewed.

Conflict of Interest Statement
The authors have no conflicts of interest to declare.

Financial Disclosure
The authors declared that this study has received no financial support.

Author Contributions
All of the authors declare that they have all participated in the design, execution, and analysis of the paper, and that they have approved the final version.

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